

CLINICAL VIGNETTE

Non-invasive cardiac imaging methods in transthyretin amyloidosis

Renata Rajtar-Salwa¹, Adam Gębka¹, Paweł Petkow-Dimitrow^{1, 2}

¹2nd Department of Cardiology and Cardiovascular Interventions, University Hospital, Krakow, Poland

²2nd Department of Cardiology, Jagiellonian University Medical College, Krakow, Poland

Two main types of amyloidosis are distinguished: abnormal transthyretin (ATTR) and light-chain amyloidosis. The gold standard for the diagnosis of cardiac amyloidosis is myocardial biopsy. Unfortunately, this examination is invasive, not easily accessible, and may fail to detect the disease if the biopsy sample is obtained from a myocardial region not infiltrated by the amyloid [1]. Thus, non-invasive imaging methods may be useful [1]. A 43-year-old man with severe heart failure (New York Heart Association class III/IV) and an initial suspicion of hypertrophic cardiomyopathy or amyloidosis was referred to our hospital for a differential diagnosis. Echocardiography (Fig. 1A) revealed a speckled pattern of the myocardium (amyloid protein is more echogenic than the surrounding myocardial tissue), symmetric left ventricular (LV) hypertrophy with infiltrated thickened valvular leaflets, enlarged left and right atria, and thickened interatrial septum. Doppler echocardiography showed a severe restrictive pattern of mitral LV filling (high E/A wave ratio [2.4], short deceleration time [110 ms], and very low early diastolic velocity of the mitral annulus [5 cm/s] in tissue Doppler imaging). Ejection fraction was 38%. At this stage, cardiac amyloidosis was considered a more likely diagnosis [1, 2]. A more advanced echocardiographic examination, i.e. the global longitudinal strain (GLS) pattern, revealed apical sparing (bull's eye view from the apex to the base; Fig. 1B). A decrease in GLS is a sensitive and specific finding that can be used to distinguish amyloidosis from other causes of LV hypertrophy. Traditional contrast-enhanced cardiac magnetic resonance (CMR) imaging showed an inconclusive finding of concentric myocardial hypertrophy but without late gadolinium enhancement (LGE). Scintigraphy with technetium-99m (99mTc) labelled 3,3-diphosphono-1,2-propanodicarboxylic acid revealed radiotracer uptake in the heart (Fig. 1C). Additional single-photon emission computed tomography and computed tomography (SPECT-CT; Fig. 1D) showed radiotracer uptake mainly in the anterior wall of the LV and interventricular septum. The uptake was lower in inferior and lateral LV walls. The patient died suddenly due to asystolic cardiac arrest recorded in continuous electrocardiogram monitoring in an Intensive Care Unit. Resuscitation was unsuccessful due to end-stage cardiac amyloidosis. In autopsy examination there were deposits of a substance that weakly stained with Congo red; however, it did not exhibit a typical apple-green amyloid glow in polarised light. Characteristic morphological manifestations of ATTR amyloidosis were focal amyloid deposits, mostly in the myocardial interstitium (pericellular). Echocardiography with additional CMR imaging can be used to diagnose cardiac amyloidosis and assess the prognosis [1].

In our case, echocardiography with modern GLS imaging was more informative than gadolinium-enhanced CMR imaging and GLS was an additional help to reach the correct diagnosis. A recent report [1] showed a GLS value equal to or less negative than -14.81% to be a predictor of all-cause mortality. Herein, the GLS value was unfavourably lower than this threshold (-12%). The crucial imaging technique, 99mTc pyrophosphate SPECT-CT, has been shown to be highly sensitive and specific (90%) for the diagnosis of ATTR amyloidosis. The results of CMR imaging were inconclusive. Traditional LGE imaging (based on inversion recovery sequence) may be limited by technical difficulties that carry a significant risk of false-negative results [3]. Myocardial T1 mapping, a novel approach to myocardial tissue imaging, can overcome the limitations of LGE imaging [1, 3]. This novel method shows high accuracy for the detection of cardiac amyloidosis and is more sensitive than LGE imaging for identifying cardiac infiltration.

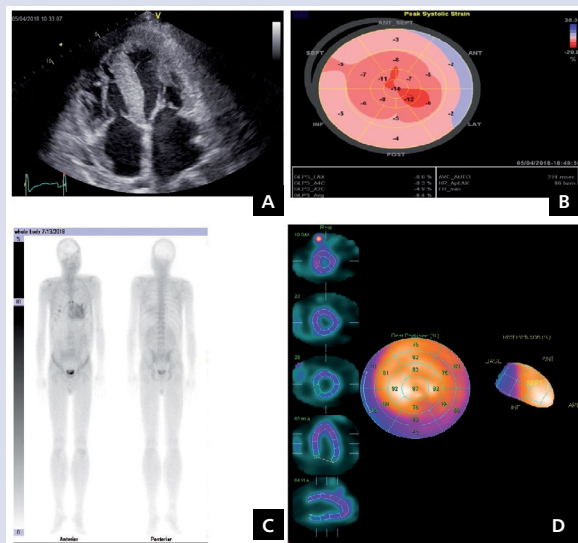


Figure 1. A. An echocardiogram showing several features typical of cardiac amyloidosis; four-chamber apical view; B. Echocardiographic reconstruction; a bull's-eye display of "polar map" of global longitudinal strain (unfavourable phenomenon, low negative percentage values); C. A whole-body scintigraphy scan showing radiotracer uptake in the heart; D. Single-photon emission computed tomography and computed tomography showing detailed localisation of radiotracer uptake

References

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Address for correspondence

Prof. Paweł Petkow-Dimitrow, 2nd Department of Cardiology and Cardiovascular Interventions, University Hospital, ul. Kopernika 17, 31–501 Kraków, Poland, tel/fax: +48 12 424 71 70/+48 12 424 71 80, e-mail: dimitrow@mp.pl

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